Pediatric Thoraco-lumbar Paraspinal Arteriovenous Fistulas along the Segmental Nerve

Diagnosis and Endovascular Treatment*

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Summary

To characterize the clinical presentation, imaging features and endovascular treatment of paraspinal non-vertebral arteriovenous fistulas along the segmental nerve.

Retrospective review was performed on the five patients identified in our database covering 1985 to 2003. All patients presented with an incidentally found continuous murmur over the upper paraspinal or parasternal regions before three years old. In four patients, the AV fistula was in the mid-thoracic level and at L3 in one. All AV fistulas were a high-flow single-hole fistula at the neural foramen with venous drainage into paraspinal and epidural veins without intradural reflux. All fistulas were endovascularly occluded in the same session as the diagnostic angiography. The fistula was occluded with detachable coils in one case and with N-butylcyanoacrylate (NBCA) with flow control in four cases. Complete occlusion of the fistula was obtained in all cases and all patients remained neurologically intact at the last follow up (average six years). Non-vertebral paraspinal arteriovenous fistula along the segmental nerve is a specific disease entity seen in children. Embolization is the first choice of treatment for this disease.

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Introduction

We retrospectively reviewed a specific subentity of paraspinal AVFs: non-vertebral paraspinal arteriovenous fistulas along the segmental nerve.

Material and Methods

From 1985 to 2003, spinal angiography with intent to treat was performed in 288 patients with spinal arteriovenous (AV) malformations. Among them, we identified five patients with non-vertebral paraspinal arteriovenous fistulas along the segmental nerve. We retrospectively reviewed the clinical presentation, imaging features, and endovascular treatment of this unique subgroup.

Results

Table 1 summarizes the data of the five patients identified. All five patients were between two and three years old at the time of diagnosis and treatment. All patients presented with an incidentally found, continuous murmur auscultated over the upper paraspinal or parasternal regions. All patients had normal neurological exams and had reached developmental milestones.

Three patients had pre-treatment magnetic resonance imaging (MRI) that showed promi-

nent flow voids in paraspinal and epidural regions suggestive of a spinal arteriovenous malformation. No intradural flow voids or T2 high signal abnormality within the spinal cord was present in any of the patients.

All patients underwent spinal angiography and endovascular embolization during the same setting under general anesthesia. In the last three cases, both transcranial motor evoked potentials (MEPs) and sensory evoked potentials (SEPs) from the four extremities were monitored and were found to be normal throughout the procedure [20].

In all cases, spinal angiography demonstrated a single-hole arteriovenous fistula directly from segmental artery branches at the level of the neural foramen into the paraspinal veins and epidural venous plexus. No pial arterial feeders or intradural draining veins were identified.

The fistula was occluded using detachable coils in one case and N-butyl-cyanoacrylate (NBCA) injection in four cases [21]. Before NBCA injection, flow through the fistula was decreased either by placing coils into the venous side in two cases or by inflating a non-detachable balloon in the feeding artery in the other two cases.

Immediate complete angiographic occlusion of the fistula was achieved in all five cases without complication. Short term follow up MRI was obtained in all cases between 1.5 and five months after the procedure. They all showed no evidence of a remaining patent arteriovenous fistula. All patients were normal in neurological and cardiovascular conditions on clinical follow-up ranged from six months to 13 years three months, average six years after the treatmement.

Table 1 **Summary of demographics, clinical presentation, imaging results, and therapy in five patients with paraspinal arteriovenous fistulas.**

Patient Number	1	2	3	4	5
Age (years) at Tx	3	3	2 1/2	3	2
Sex	Male	Female	Male	Male	Male
Presentation	Continuous murmur	Continuous murmur	Continuous murmur	Continuous murmur	Continuous murmur
Echocardiogram	Mild cardiomegaly	No cardiomegaly	No cardiomegaly	No cardiomegaly	Right atrium and ventricular enlargement
MRI performed	Yes	No	No	Yes	Yes
Level of arterial feeder	Left L3	Left T6	Left T7	Right T7	Left T5
Fistula holes	Single hole	Single hole	Single hole	Single hole	Single hole
Venous drainage	Epidural paraspinal	Epidural paraspinal	Epidural paraspinal	Epidural azygos	Epidural paraspinal asygos hemiazygos
Material used for embolization	Coils	NBCA with balloon flow control	Coils: NBCA	NBCA with balloon flow control	Coils: NBCA
Occlusion	Complete	Complete	Complete	Complete	Complete
MRI follow up	4.5M	5M	1.5M	2.5M	3.5 M
Clinical Follow-up	9 yrs, 7 months	2 yrs, 2 months	13 yrs, 3 months	1 yr	6 months
Clinical Outcome	Normal	Normal	Normal	Normal	Normal

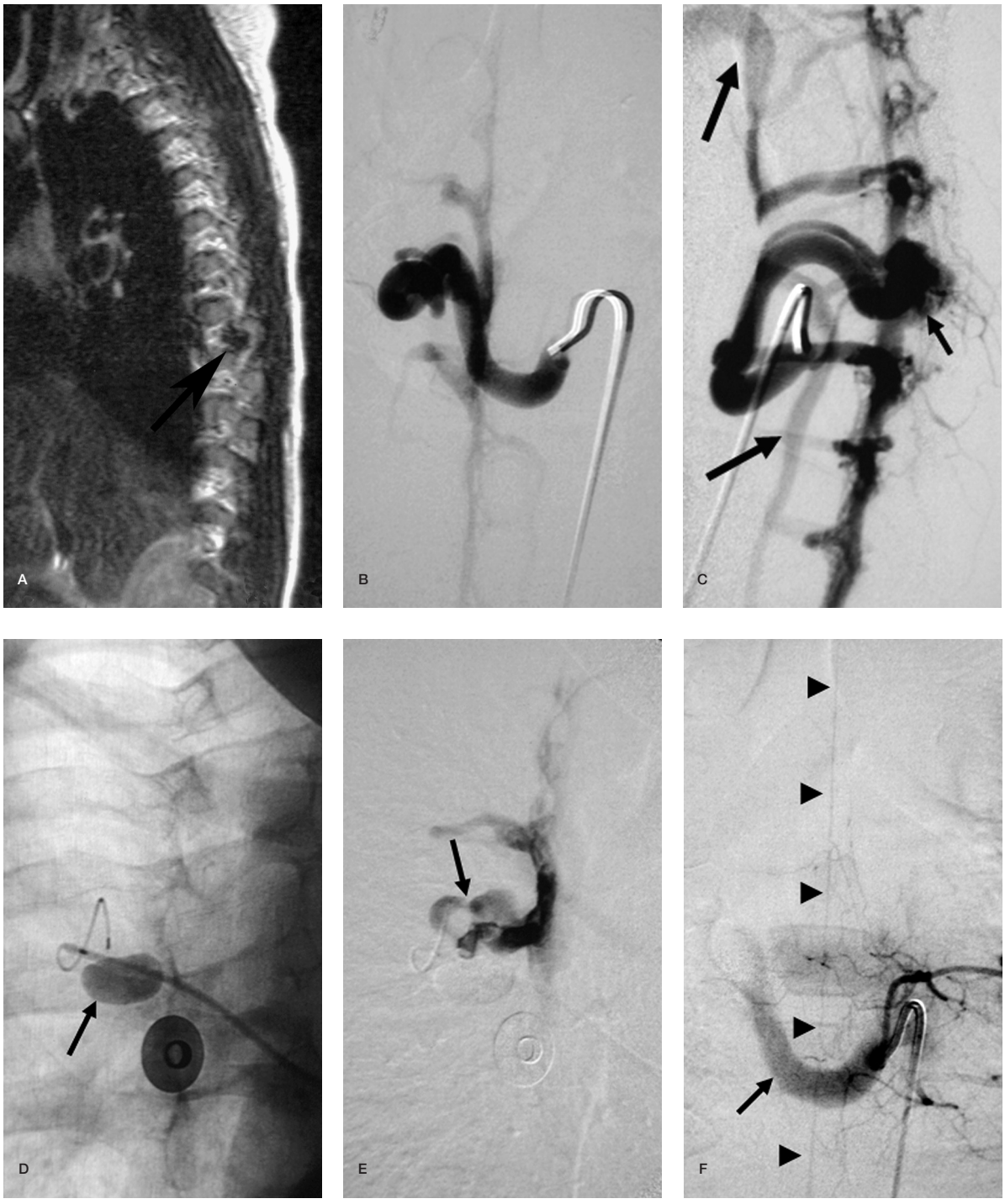

Figure 1 A) Lateral T1-weighted MRI of thoracic spine shows a low signal intensity lesion in a paraspinal location adjacent to a nerve root (arrow). B) AP view of right T7 intercostal artery angiogram demonstrating a high flow single hole fistula along the nerve root draining to the epidural venous plexus. C) Lateral view of right T7 intercostal artery angiogram demonstrating a single hole fistula along the nerve root (arrow) draining to the epidural venous plexus. There is opacification of venous drainage to azygos hemiazygos system (large arrows). D,E) Non-subtracted (D) and subtracted (E) superselective angiogram from the microcatheter placed near the fistula site (arrow in E) through the double lumen balloon catheter which was inflated to decrease the flow through the fistula (arrow in D). F) Left T7 intercostal artery angiogram after embolization demonstrating preservation of the anterior spinal artery (arrow heads). There is stagnation of contrast in the proximal right T7 intercostal artery which previously supplied the fistula (arrow).

Illustrative Cases

Case 1

This 3-year old boy was brought to medical attention when a loud murmur was detected over his upper back by his mother. At this time, the child was asymptomatic and meeting all of his developmental milestones.

An echocardiogram showed a normal heart. The child was presumptively diagnosed with a pulmonary arteriovenous fistula and referred to us for embolization. Thoracic MRI revealed a large flow void in the paraspinal area in the mid-thoracic level on the right (figure 1A).

Spinal angiography showed a high flow arteriovenous fistula along the right T7 nerve supplied by the markedly hypertrophied intercostal artery and drained through the epidural venous plexus into the azygous vein (figure 1B,C). With the assistance of a balloon-catheter used to achieve proximal flow control angiography, superselective angiography revealed a single hole fistula into the paraspinal vein (figure 1D,E). Embolization using a 40% NBCA mixed with Ethiodol resulted in complete occlusion of the fistula with preservation of the anterior spinal artery (figure 1F). The child awoke without neurologic deficit. Follow up MRI at 2.5 month showed no evidence of a patent fistula. At one year, the child is developing normally and is asymptomatic. The murmur has disappeared.

Discussion

Among spinal vascular lesions, paraspinal malformations are the least recognized entities. They can be detected incidentally or with venous congestive myelopathy or with compressive myelopathy [1,4-6,10]. They can also present with a subcutaneous pulsating mass in the paravertebral musculature, subarachnoid haemorrhage, or spinal canal deformity [8].

Fistulas along the segmental nerve can be a solitary lesion or associated with a spinal cord AVM as a part of metameric malformation. The most recognized segmental nerve fistula is the spontaneous vertebral artery fistula, typically seen at the C1-C2 or C7 vertebral levels, which are often found in children. As a rare variation of segmental nerve fistulas, a fistula can be supplied by a single intercostal or lumbar artery. There are few reports on non-vertebral paraspinal arteriovenous fistulas along the segmental nerve in the literature, and they are not well-recognized as a distinct sub-entity of the segmental nerve fistulas.

By retrospective analysis of 288 patients with spinal AVMs, we found five non-vertebral paraspinal AVFs along the segmental nerve and discovered their remarkable homogeneity of clinical presentation and angiographic appearance.

They were all diagnosed at two to three years of age with an incidentally discovered continuous murmur over the paraspinal or parasternal area. The AV fistula was located in the midthoracic level in four patients, and at L3 in one patient. All lesions were high-flow, single-hole fistulas located at the level of the neural foramen.

The venous drainage was only into paraspinal and epidural veins without reflux into the perimedullary veins. No metameric lesions were seen. Despite the presence of a high-flow AVF, none of our patients had clinically symptomatic cardiac failure. This may be secondary to relatively high resistance in the epidural venous plexus. Angiographic studies of the two patients who had mild cardiomegaly showed early visualization of the azygos/hemiazygos veins draining into the superior/inferior vena cava and then to the right atrium.

The lack of reflux into perimedullary veins and consequent lack of signs and symptoms of spinal venous hypertension likely explain the normal neurological exam documented in our five patients. In the few pediatric cases which appear to be paraspinal segmental nerve fistulas that have been reported in the literature, no perimedullary venous drainage has been described [8,9]. Therefore, it appears rare that non-vertebral paraspinal AVFs along the segmental nerve develop intramedullary venous drainage. This is probably because the fistula is distal to the antireflux mechanism of the radicular vein at the site of dural penetration [7,12,13].

This is opposed to SDAVFs which are located inside the dural membrane at the level of the anti-reflux mechanism where rich axial and longitudinal anastomoses exist to the epidural venous plexus.

Although we do not know their natural history, we think that non-vertebral paraspinal AVFs along the segmental nerve can eventually lead to venous congestion of the spinal cord. Myelopathy may be caused by either pure venous hypertension of the epidural plexus without intradural reflux or by the development of

intradural reflux through the radicular vein draining into the epidural venous plexus. Such venous congestion may result because long-standing high-flow fistulas can develop venous stenosis, pouches, or thrombosis. A few cases have been reported of extensive epidural AVFs or vertebral fistulas with intradural reflux of the venous drainage which presented with subarachnoid haemorrhage or progressive neurological deficits due to venous hypertension of the spinal cord [3,5,6,14]. The risk of developing high output cardiac failure from a longstanding AVF may also exist. Development of neurological symptoms due to mechanical compression of the radicular nerve or the spinal cord is also possible as reported in vertebral fistulas.

We believe that endovascular embolization is the treatment of choice for this diesase. Before endovascular treatment, it is mandatory to analyze the normal vascular anatomy of the spinal cord as well as the lesion itself. On the analysis of the vascular anatomy of the lesion itself, it may be difficult to visualize the exact fistula site by conventional angiography because of high flow through the fistula and overlapping venous structures.

We recently started using three-dimensional (3D) angiography to analyze the vascular anatomy of spinal vascular lesions [11].

The obliteration of high-flow AV fistulas can be achieved with a liquid embolic agent such as NBCA, coils or detachable balloons [2] or combination of these materials. NBCA as a permanent embolic agent offers an immediate and durable occlusion for high-flow AV fistulas but some kind of flow control method during embolization to prevent migration of the embolization material into the venous system. In cases three and five, coils were placed in the venous side to decrease the flow through the fistula. In cases two and four, a proximal temporary balloon occlusion provided arrest of flow during NBCA injection.

Conclusions

Non-vertebral paraspinal arteriovenous fistula along the segmental nerve is a specific subentity of paraspinal AVMs. They often present in childhood as incidentally discovered bruit and consist of a high-flow single-hole fistula from a segmental thoracic or lumber artery to the epidural venous plexus. This lesion should be treated to avoid potential neurologic or cardiac complications due to a long-standing high flow fistula. Endovascular embolization is a safe and effective treatment modality for this condition and should be the first treatment option.

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